CASE REPORT

Cutaneous Vasculitis in a Patient with Dermatomyositis without Muscle Involvement

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A 74-year-old female patient with cutaneous ulcerations and typical dermatomyositis (DM) skin rash had no muscle disease for a 1-year and 5 months period. Histological examination of the skin ulceration indicated vascular occlusion without cellular infiltration. Cutaneous ulceration is a very rare manifestation of adult-onset DM patients without inflammatory myopathy. (Internal Medicine 33: 809–812, 1994)

Key words: skin ulceration, amyopathic dermatomyositis

Introduction

Dermatomyositis (DM) is a connective tissue disorder characterized by prominent cutaneous features and inflammatory myopathy. A DM patient may not necessarily have both cutaneous and muscle disease at the initial presentation. Cutaneous manifestation of DM frequently precedes the development of muscle disease. The absence of muscle disease in patients with the classical skin rash of DM persisting over a period of 10 years has been reported (1). Amyopathic DM is the term used to describe patients with typical cutaneous manifestation of DM with no or minimal muscle disease.

Cutaneous vasculitis characterized by periungual infarcts and digital ulcerations has been noted frequently in child-onset DM. However, its occurrence in adult-onset DM is rare.

The patient in this study had the typical skin rash of DM without apparent muscle disease; subsequently digital and foot ulcerations developed. The rare association of skin ulceration and amyopathic DM in adults is discussed.

Case Report

A 74-year-old Japanese female was first admitted to Kitasato University Hospital in April 1993 with a two-month history of fatigue, swelling of the fingers and scaly and erythematous eruptions on proximal interphalangeal joints, metacarpophalangeal joints, elbows and knees. She had no past or family history of collagen disease nor had she experienced Raynaud's phenomenon or arthralgia.

Examination indicated an edematous and very faint purplish erythematous eruption on proximal interphalangeal (PIP) joints, metacarpophalangeal (MCP) joints and extensor surfaces of the elbows (Fig. 1) and knees. A faint purple, scaly and macular eruption was noted on the clavicular and V-area of anterior part of the chest. There was no indication of muscle weakness. Levels of muscle enzymes such as serum aspartate aminotransferase, creatine kinase and aldolase were normal. Serum lactic dehydrogenase was 551 IU/l (normal range; 180–350) on admission, but it decreased to normal ranges without treatment. The results of an electromyogram were negative. A muscle biopsy was performed from right biceps brachii muscle. Muscle specimens did not exhibit atrophy, degeneration or fibrosis of the muscle fiber. Distribution of type I and type II muscle fibers were indicated to be normal. Infiltration of inflammatory cells and vasculitis were absent in the specimens. Histological examination of skin specimen obtained from the fingers and elbows indicated hyperkeratosis of epidermis, thickness and edematous changes in collagenous connective tissue of the dermis, basal keratinocyte liquefaction degeneration and mild infiltration of mononuclear cells around small blood vessels (Fig. 2). These findings were consistent with the features of skin eruption of DM. A diagnosis of DM without prominent muscle disease was made. The patient was treated with prednisolone 20 mg/day; the skin eruptions showed good response.

At one month following discharge from the hospital, a skin ulceration of the right fifth finger and left sole developed during prednisolone treatment of 17.5 mg/day. The right fifth finger then became edematous and erythematous with subsequent spreading over her right forearm. In August 1993, she was admitted again. She had Gottron's papules on PIP joints, MCP...
joints and typical rash of DM on the elbows and knees. Periungual erythema, multiple nail fold thrombi on fingers and scattered papular erythema on the fingers and soles were noted. A small digital ulceration 3 mm in diameter on the dorsum of the right fifth PIP joint, deeper skin ulcerations 12 mm in diameter on the left sole and 6 mm in diameter on the left fifth toe were detected (Fig. 3). The tip of the left fourth toe was gangrenous.

Swelling and tenderness of the right arm due to cellulitis were evident. No sclerodactyly or muscle weakness could be detected. Muscle enzymes such as aspartate aminotransferase, creatine kinase, aldolase and lactic dehydrogenase showed normal range levels. The erythrocyte sedimentation rate (ESR) had increased to 72 mm/h and C-reactive protein (CRP) was 5,048 μg/dl. Serum IgG and IgA were increased to 1,880 mg/dl and 514 mg/dl, respectively. Antinuclear antibody was positive at ×80 with a speckled and homogenous pattern, but the antibody to extractable nuclear antigen was negative. Antineutrophil cytoplasmic antibody (ANCA) and anticardiolipin antibody were not detected. The results of the electromyogram were also negative. To analyze for occult malignancy, X-ray examination, abdominal echogram, chest and abdominal CT scan, gallium radioisotope scanning, gastrofiberscope and barium enema were performed. No associated malignancy was detected. Pathological examination of skin biopsy specimens from the right sole demonstrated dilatation of vessels in the dermis with endothelial hyperplasia, vascular occlusion due to intraluminal thrombi; there was no cellular infiltration around the vessels (Fig. 4). Immuno-histochemical examination disclosed the deposition of IgG, IgA, IgM, C3, C4, C1q and fibrinogen on the vessels.

The patient was treated with heparin (10,000 U/day) and lipo-PGE1 (10 μg/day), but without significant effect. Methylprednisolone pulse therapy (500 mg/day for 3 days) was followed by oral administration prednisolone 40 mg/day and warfarin. The skin ulcerations were resistant to the therapy, but significant improvement was noted at 4 months.
Skin Ulceration in Amyopathic DM

Discussion

Dermatomyositis is the designation for patients who have both inflammatory myopathy and typical skin eruption. The onset of a skin disorder does not necessarily occur at the time of muscle involvement. Bohan et al found that 93% of 45 patients with DM had a skin rash at the time of presentation but muscle weakness was present in only 53% (2). Rockerbie et al observed that skin rash preceded muscle weakness in 56% of DM patients. Skin rash preceded muscle disease by more than 1.75 years in 12% of this group (3). Krain reported six cases of DM showing typical cutaneous changes without apparent muscle involvement at the time of presentation (1). All these patients eventually developed inflammatory myopathy. It is thus apparent that skin rash of DM commonly precedes muscle involvement.

Pearson demonstrated five cases of DM with typical rash but no indication of muscle weakness (4). One of these patients had rash for 13 years without muscle weakness. Thus, amyopathic dermatomyositis is a designation applicable to patients showing typical cutaneous manifestation of DM with no or minimal muscle disease. Euwer and Sontheimer reported six cases of amyopathic dermatomyositis in whom the skin manifestations occurred two years prior to muscle disorder; they maintained that this term was only a provisional diagnosis for some form of DM (5). Four cases of DM demonstrated only skin eruption after 4 to 11 years; these may be true cases of amyopathic dermatomyositis (6). Thus, not only does skin rash frequently precede muscle involvement, but skin manifestations alone are expressed in some patients.

Clinical evidence for muscle disease was not found until 1 year and 5 months after the onset of the typical skin rash of DM in the present patient. The systemic administration of glucocorticoid hormone may have caused delayed development of inflammatory myopathy in this case. The patient developed severe cutaneous vasculitis during therapy, and thus may belong to a subset of DM showing cutaneous manifestation of DM with no muscle disease. There is no established criteria for amyopathic dermatomyositis. Therefore, the patient may be tentatively diagnosed as amyopathic dermatomyositis. Muscle involvement should be evaluated carefully in this case to determine the applicability of this term. The limited number of amyopathic DM patients makes it difficult to confirm the clinical characteristics of the disease. Stonecipher et al reported four cases of amyopathic DM with skin manifestation alone showing good prognosis. However, two cases with malignancy were found among nine amyopathic DM patients in whom developed myositis subsequently (6). Fudman and Schnitzer also demonstrated three patients with malignancy and two patients with severe interstitial pneumonitis among seven DM patients who had cutaneous changes and muscle involvement but normal creatine kinase levels (7). Two patients with amyopathic DM in Japan have been noted to exhibit fatal interstitial pneumonitis (8). Thus, some patients with amyopathic DM have been demonstrated to have a poor prognosis.

Systemic vascular injury is the characteristic pathognomonic feature of child-onset DM. Cutaneous vasculitic ulceration has been observed frequently in one-fourth of child-onset DM patients, but it is rarely noted in adult-onset DM. The clinical features of adult-onset DM showing cutaneous vasculitis have not been well characterized. Feldman et al observed seven cases (9.2%) with cutaneous vasculitis. They detected subcutaneous nodules in two, periungual infarcts in three and digital ulceration in two, of 76 patients with adult-onset DM (9). Increased association of cutaneous vasculitis and malignancy was noted in their work. In the Japanese literature, four cases of DM associated with cutaneous ulcerations are reported (10–13). However, associated malignancy was not found in these cases. No patient with adult-onset DM without muscle involvement, such as amyopathic dermatomyositis, manifesting cutaneous ulceration as a feature of skin disease, has been reported.

Various pathological changes in cutaneous vessels have been noted in childhood DM (14). Endothelial swelling, vascular occlusion and infarction and necrotizing vasculitis associated with cellular infiltration have been shown. The deposition of immunoglobulin, complement and fibrin on vessel walls has also been observed. In the present case and in three reported cases of adult-onset DM with cutaneous ulceration, vascular occlusion or lymphocytic infiltration around subcutaneous vessels were demonstrated.

The present patient was treated with methylprednisolone pulse therapy followed by prednisolone and anti-coagulant therapy. Although skin ulceration was quite resistant to therapy, significant improvement was noted at 4 months. Cutaneous ulceration was reported resistant to steroid therapy in one report (9). Thus, cutaneous ulceration due to vasculopathy in DM patients should be treated carefully due to the resistance to the therapy.

References

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